Sarcoidosis

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Disclosure

• No financial conflict of interest
What is Sarcoidosis?

- A chronic inflammatory disease which can affect any organ of body but most commonly lungs.
What is Sarcoidosis?

• Characterized by overreaction of immune system which results in the build up of several small clusters of inflammatory cells called “Granulomas” in the affected organs.
What is Sarcoidosis?

- Formation of too many of these clusters may interfere with organ structure and function.
Possible causes

• No one knows exactly what causes sarcoidosis

• Thought to be resulting from one or more exposures to “trigger factors” in people with certain genetic make up.
Possible causes

Suspected triggers

– bacteria,
– viruses,
– fungi
– chemicals

• These triggers are usually harmless in most people, but may irritate the immune systems of people who are at genetic risk for developing sarcoidosis.
Risk Factors

• Affects people worldwide, of any age, race, gender

• In US, most common and severe in African Americans and European “particularly Scandinavian” descent.

• Age: 20 - 40 yrs of age

• Gender: Women > men
Associated Risk Factors

• Environmental exposure
  – inorganic particles, insecticides, and mold

• Occupational exposure
  – U.S. Navy, metalworking, firefighting, and the handling of building supplies

• Fire fighters and rescue workers involved in 2001 WTC disaster
Genetic Factors

- Risk higher if close family members have sarcoidosis
Symptoms

- Many people are often asymptomatic, detected incidentally on chest X-ray done during routine physical exam.

- Symptomatic people can have variable signs and symptoms depending on the organs affected.
Symptoms

Common symptoms:
- shortness of breath,
- wheezing,
- chronic cough
- Chest pain
Other Symptoms

- Fatigue
- Weight loss
- Night sweats
- Skin rashes
- Eye symptoms
- Irregular heart beat
- Leg swelling
- Joint pain, swelling
- Muscle soreness
- Weakness & Numbness
- Enlarged Lymph nodes
- High blood or urine calcium level
- Kidney stones, Renal failure
Diagnosis

• Sarcoidosis is a diagnosis of exclusion

• There is no objective test which can easily diagnose sarcoidosis.

• Numerous exams and tests are required to confirm the diagnosis and to decide the best treatment options.
Diagnosis

- Chest X-ray
- CT scan
- Lung function test
- Biopsy of the affected organs (Lungs, lymph nodes, skin, bone, liver)
- Blood tests (Blood cell count, liver test, renal function test, blood and urine calcium, ACE level)
- MRI
- PET scan
- Cardiac tests (EKG, Echo, heart rhythm monitoring)
Clinical course

• In many cases of sarcoidosis, no treatment is necessary and sarcoidosis may go away in 2-5 years without medical treatment.

• The disease may never reappear or may reappear (in ~ 5%) later in life.

• In other patients, the disease is progressive, causing scarring in affected organs and requiring ongoing treatment.
Treatment

• Treatment is not indicated in asymptomatic patients

• These patients are monitored closely for worsening

• Treatment is started when symptoms are severe enough to impair quality of life or when there are danger organ damage or life.
Medications

- **Corticosteroids: Prednisone**
  - First line treatment
  - Reduces inflammation
  - Starting dose 20-40 mg daily
  - Relieves symptoms in 1-3 months
  - Then taper prednisone slowly to the lowest dose which will keep symptoms under control and cause less side effects
  - Stay on this dose for another 6 months then taper it off
  - Common Side effects: weight gain, insomnia, mood swings, acne, high blood sugar, osteoporosis, risk of infections
Medications

• Immune system suppressant medication:
  – Methotrexate, Azathioprine, Leflunomide, Cellcept
  – 2nd line treatment
  – Indicated when prednisone is not effective or causing significant side effects
  – Can take up to 6 months to show its effects
  – Side effects: Liver, kidney, bone marrow toxicities, malignancies
  – Blood tests are done every 1-3 months to monitor side effects
Medications

• Antimalarial drugs:
  – Hydroxychloroquine (Plaquenil)
  – Effective in skin symptoms or a high level of calcium
  – Side effects: Nausea, vision problem
  – Needs eye exam every 6 months
Medications

• Biological agent (Anti-TNF)
  – Infliximab (Remicade) IV
  – Adalimumab (Humira) SQ
  – Third line treatment
  – Side effects: Risk of infections, liver toxicities, lupus like illness, cancers
Treatment

• Sarcoidosis is often treated with the help of a multidisciplinary team of health care professionals who specialize in the treatment of the lungs, heart, brain, kidneys, liver, eyes, and skin.

• At specialized medical centers, these health care providers work as a team to develop a comprehensive treatment plan.
Follow up

- Asymptomatic patients
  - Initially every 6 months, then annually for 3-5 years

- Symptomatic patients
  - Every 3-6 months

- Follow up test:
  - Blood tests and PFT every 3-6 months
  - Chest X-ray every 6-12 months
Complications

- Fibrosis of lungs
- Cavities in lungs with fungal ball formation
- Pulmonary hypertension
- Side effects of medications
- Fatigue, depression, small fiber neuropathy
Take home points

• A chronic inflammatory disease characterized by formation of clusters inflammatory cells “granuloma”

• Exact cause unknown but is thought to be due to overreaction of immune system in response to exposure to triggers in people with certain genetic make up

• Has been associated with exposure to inorganic particles, insecticides, molds, certain occupations line navy, firefighters, metalworking,
Take home points

- More common and severe in AA
- Risk increases if close relatives have sarcoidosis
- Can affect any organ but most common in lungs
- Biopsy of affected organ is needed for diagnosis
- Many patients are asymptomatic and may go away in 2-5 years without medical treatment
- Common symptoms are respiratory followed skin, eye
- Treatment is indicated only in symptomatic patients or if affecting, heart, brain
- First line treatment: Prednisone
Thank you

• Questions?

Question

What is the definition of Sarcoidosis?